cCMV: Essential Knowledge for Audiologists

Presenters: Deborah Flynn, AuD, Alissa Nickerson, AuD, and Wendy Steuerwald, AuD

- [Christy] It is my pleasure to welcome a wonderful group of audiologists from Phoenix Children's Hospital present to us today on cCMV and the essential knowledge that we need as an audiologist. My pleasure to welcome Dr. Wendy Steuerwald. Over to you, Wendy.

- Thank you so much for that great introduction. Here are our disclosures, I'm not going to read them, but they're here if you wanna look at them. Our learning outcomes for the day are, after this course, you'll be able to explain the difference between symptomatic and asymptomatic cCMV, list three ways to reduce transmission of CMV, and explain the audiology management for patients with cCMV. So I'm so excited to share with you guys that Katie Hobbs, the governor of Arizona, has made a public proclamation that June is CSUN CMV Awareness month in the state of Arizona, so, hooray.

What is cytomegalovirus, otherwise known as CMV? CMV is a common herpes virus, and what this means is that once you get it, you have it for the rest of your life. Sometimes it's active, sometimes it's inactive, but it's in your system. It's typically harmless when acquired by children or adults. Children or adults who get it will have common cold-like symptoms. It's common in kids, especially those in daycare or around other kids. What is cCMV, which is congenital cytomegalovirus, and this is CMV which is acquired by the infant while in utero. So their mother contracts CMV and it passes through the placenta to the infant, which is where all the issues occur because the baby has cCMV while their organs are developing and while they're growing. cCMV is the most common infectious cause of birth defects in the United States, impacting one of every 200 births. That's a lot. And of great importance to us audiologists, it's the leading cause of non-genetic childhood onset sensory neural hearing loss. Infants with CMV can be symptomatic or asymptomatic, and CMV testing is recommended to be performed before the child is 21 days old to differentiate CMV from cCMV. If you wait much longer than that, there's no way to know did the child acquire CMV before birth

or after birth. CMV is complicated and it's complicated by a lot of factors, including lack of community and medical awareness.

As I was saying before the session started, this is one of those diseases which is everywhere, but it's not talked about, and most people don't know a lot about it. Kathleen Muldoon is a researcher and she presented CMV 101, which was the first presentation in this CMV Awareness series on AudiologyOnline. She's also done many research papers on CMV awareness, so if you wanna learn more about that, you can just look for her name and Google, and they will come up. To increase awareness, The American Academy of Audiology released a position statement in March of this year. So that's available on AudiologyOnline, or you can look it up in Google Scholar. The lead author is Maggie Kettler. There are many states, including Arizona, which have websites to discuss CMV and to raise awareness. In Arizona, we have Stop CMV AZ, and a Spanish version, which is Alto CMV AZ.

There's also the National CMV Foundation. There are many states that have proclamations that June is cCMV Awareness Month, including Arizona and Ohio. There's also state legislation from quite a few states regarding cCMV. I think we all are excited about the impact that Minnesota's universal cCMV screening is going to have on the children of Minnesota and on the rest of the country. Utah has a fabulous public awareness program, and if you want to know more about that, you can check out Stephanie McVicar's presentation, which she did live last week, "Congenital CMV Advocacy and Legislation." Also, I've been noticing a lot more cCMV presentations at various conferences. There was one by SENTAC a couple weeks ago, ACIA last week had several cCMV talks and of course AudiologyOnline. Further complications are, there's a lack of universal screening as we all know, and also there's variations in the way to screen. We can screen for CMV through buccal swab, which is just a simple cheek swab, urine, and by blood spot. And all of these ways have different sensitivity and there's also variations in models of who to screen.

Some states such as Minnesota have universal screening. Other states just screen babies who don't pass the newborn hearing screen and still other states have a hearing targeted screening, which is if the child has hearing loss and something else, they'll go ahead and screen. And something that I just learned, you all may already know this, but I had no idea, but there's differences in the tests that labs run to assess whether or not conditions such as CMV are positive. Now, some states use blood spots very creatively in that if a child has a progressive hearing loss, they can check the blood spot which was taken at birth to see if CMV was positive in the blood. Fascinating, fascinating. Something I learned yesterday from Dr. Padron's presentation, he did the Pathophysiology Diagnostic and Treatment session. One of the things that he talked about was if a mother contracts CMV while they're pregnant, they're able to test for it and treat it. But some places are like, yes, we should do this. Others are like, no, not a good idea. So there's lots of controversy over that.

As I mentioned earlier, there's also inconsistent presentation. Some children are born symptomatic, some are asymptomatic. We did a pilot study here at St. Joe's Hospital in Phoenix, Arizona, and if you want to learn more about it, check out the Hearing Loss in cCMV presentation, which doctors Page and Bascuti did a few weeks ago. But just to give you a little synopsis, we tested 250 infants from the Well Baby Nursery and found two of them cCMV positive by buccal swab. But then we did a urine confirmation and only one was found positive. So a lot to think about with cCMV. Also, there's a great range of severity. Children who have it can have mild symptoms up to very severe symptoms. So, symptomatic versus asymptomatic. 90% of the babies with cCMV don't have any symptoms, 10% of them have symptoms. Now, I've included here a list of common symptoms. This doesn't mean that all babies who have symptomatic CMV are going to have all of these symptoms. Some may have one, some may have two, some may have a whole lot of them. Again, a great range in symptoms. One of the problems is because many infants are asymptomatic at birth,

and cCMV is not universally screened for in the US, these infections can go undiagnosed. So you're probably wondering, I wanna have a baby. How do I keep myself safe?

Well, there's no vaccine for cCMV yet, but vaccines are in development. Moderna is actually running clinical trials right now, and if you'd like to participate in them, just use the web link I put there and go and sign up. Merck is also doing cCMV vaccine development. So we have two good shots here of getting a vaccine for CMV quickly. Now some things you need to think about is that the shedding of the virus can occur with or without signs or symptoms. Adults shed the virus for less duration compared to children, typically less than six months. Those of us who are in close contact to children under two years old are especially susceptible because the virus is released in the saliva and urine for quite a long time. And a study looked at kids at 18 months of age and those who had cCMV, they were all shedding in their urine, but the same children, only 24% were shedding in their saliva. So if you have a little one at home or you babysit on the side, make sure you wash your hands thoroughly after you change diapers. Shedding peaks at one to two years of age, suggesting very young children pose a risk of transmission.

Contact precautions are recommended for healthcare workers exposed to CMV. How to reduce your risk of CMV infection. Saliva content is the most likely root for transmission of pregnant women. So this is what you need to think about. Don't share anything that goes in a child's mouth. Don't put it in your mouth. Everyone gets their own cup. Don't clean the pacifier by putting it in your mouth. Kiss kids on the top of their head or the cheek, not on the mouth. Wash your hands all the time using hot soapy water. Say the ABCs, do it for the full length of time. And thinking about the clinic, make sure you clean toys, countertops, other clinic areas very, very well. Remember, CMV can live on plastic for up to 15 minutes, so clean everything very well. Disinfect equipment and furniture, use disposable equipment. If a child sneezes on the

tissue box or anything that can't be cleaned, give it to them or throw it away. At this point I'm going to hand off to my colleague, Alissa Nickerson.

- All right, thank you so much. I am a clinical audiologist at Phoenix Children's Hospital. And so this topic is really near and dear to my heart because we see kids routinely who have cCMV in our clinic. And the first thing I'd like to talk with you about are audiology considerations for patients with CMV. As Wendy mentioned, some children are symptomatic at birth, meaning that they're showing other signs and symptoms aside from hearing loss. And so that might be liver problems or in utero growth restriction. Other babies show no symptoms at birth and they appear otherwise healthy. These kiddos who have symptomatic CMV, they're the ones who are more likely to have hearing loss, although both groups can be impacted. And so studies have shown about 22% to 65% of symptomatic babies will develop hearing loss and only 6% to 23% of those asymptomatic babies will develop hearing loss.

If you're an audiologist and you're looking to learn more about cCMV and hearing loss, I definitely would recommend that you look at "Hearing Trajectory in Children with Congenital CMV," and that can be found in the Journal of Otolaryngology, Head and Neck Surgery. In this article, they talk about hearing findings for kids with CMV. What we know is that hearing loss can be unilateral, impacting one ear. It can be bilateral, impacting both ears. It can be congenital, meaning it's present at birth. It can occur later in life, and in all situations it can progress. One thing to think about is that when hearing loss is present congenitally, when it's found at birth, it's likely to progress to severe or profound by the time the child is age five. And also when hearing loss is unilateral at birth, by the time that the child is 18, it could progress into the other ear. And so I think that's a really important thing to think about as an audiologist when we're counseling patients, even at birth during diagnostic ABR studies. A mild hearing loss at birth for a child with CMV may not always be a mild hearing loss, and it certainly could progress in one or both ears. And that may impact how the families proceed with

their decision making. It may impact their communication modes, it may impact their school placement, whether or not they're involved in early interventions. And then of course it's gonna impact what kind of devices we're selecting with these families to best help them reach their potential.

Late onset hearing loss can occur in about 30 to 50% of cases. And so what's interesting to think about here is that if a child is CMV positive, but they have a normal ABR study at birth, that doesn't mean that they won't develop hearing loss later on. And routine surveillance of these kids is really important. Because late onset hearing loss can occur and progressive hearing loss can occur, we want to be sure to keep an eye on these kids. We know that as children get older, the hearing loss tends to be more severe because of these reasons, and that the newborn hearing screening program is certainly not gonna detect all these CMV patients who will develop hearing loss. It's estimated that less than half of children with CMV related hearing loss will be flagged through that newborn hearing screening program. And I think that's further justification for the need for CMV testing at birth.

Some studies have shown that in children who have normal hearing at birth, hearing loss can actually occur even within three months of birth. And that's a pretty quick turnaround in my opinion. Okay, audiology testing schedule for patients with CMV. Up until very recently, there were many suggestions for how often we should be testing kids with CMV in the literature. As Dr. Steuerwald mentioned, the American Academy of Audiology just very recently put out a position statement for CMV and in that position statement, they endorse model two. And so with this model, we wanna be sure to be doing a diagnostic test for hearing by three months of age for patients with CMV. And then we wanna be testing every three to six months in that first year of life bi-annually, meaning twice a year until age three, and then annually, testing once a year until age six. Now keep in mind that this is just a recommendation. If a family comes to you and they say, man, I'm concerned that hearing loss might be progressing, or my child's

speech is seeming more atypical, that might be justification to see this child in sooner, to test them more frequently, because we know that hearing loss can progress and it can progress rapidly.

In the AAA position statement, you'll see their justification for why this model is recommended. But really the goal is to capture these kids who are gonna have hearing loss early on. The earlier we find the hearing loss, the sooner early interventions can be pursued. And we know that kids with CMV may have poor receptive language. And so it's not just audiology that we're thinking about, but we're thinking about speech and language development. If these kids need speech and language therapy or rehabilitation, we wanna start that as soon as we can so that they can reach their full potentials. And lastly, I won't touch too much on this 'cause we'll talk about it in future slides, but when we're identifying these hearing losses, we want to be considering the fact that they could progress and be sure to choose devices that are going to grow with the children as they get older. And next up we'll talk about special considerations for assessment.

Progression of hearing loss is common and so is a very rapid progression of hearing loss. How is that gonna impact our assessment as audiologists? Of course we wanna be testing per the schedule, but maybe testing sooner if the family has concerns. Because hearing loss can progress, that may change what devices we select for children. If a child has a mild hearing loss, sometimes families might be inclined to choose a small, low gain hearing aid for aesthetic purposes, but that may not be the best long-term option for a child with CMV who may have progressive hearing loss. And so counseling the family that there's other options that might accommodate a change in hearing down the road, that's really, really important because we're not wanting to create additional burdens for families who are already navigating the healthcare system, specifically a financial burden of having to purchase new devices if the hearing loss does progress. Another thing that we wanna think about when we're

counseling families is that although a hearing loss may be mild or moderate at birth, it could progress certainly to a severe profound range. And I think educating families so they know this early on is important. It's important so that they can plan for the future and also so they really know what the future might look like for their baby. Earlier intervention is always better. And if they're a candidate for a cochlear implant, of course we'd wanna do that as soon as we could. We know that symptomatic babies are more likely to have severe consequences, and because of that, there's often many healthcare providers involved in the care of these babies. We're gonna be taking a multidisciplinary approach.

And here at Phoenix Children's Hospital where we see this the most is maybe a patient is developmentally behind, maybe they're not able to participate in behavioral assessment, we might opt to do an ABR study under anesthesia. And when we can coordinate with other disciplines, that's certainly in the best interest of the patient and their family. We know that cCMV can impact vision. So a dual sensory impairment is possible. That's gonna impact our testing as audiologists, absolutely, especially if we were planning to do visual reinforcement audiometry. So we need to be looking at the child as a whole and choosing techniques that are gonna get the best results for our patients. And then lastly on this slide is we know that cCMV can impact the vestibular system. Studies have shown that even in the presence of typical or normal hearing, children can have vestibular impairments. And so we want to be referring for vestibular evaluations when they're warranted and talking to the healthcare team so that appropriate referrals can be made.

All right, as was mentioned previously, here in Arizona, we do not have a universal cCMV screening program in our state. We'd love if that was different and we hope to change that in the future. But what we did do previously was a pilot study. And so that pilot study, we were starting to think about what would happen if universal screening was in place here in Arizona. And what we know is that we need guidelines in place

and we need to have a testing schedule for these patients. And in doing so, we looked back at our patients here at Phoenix Children's that do have CMV. And I wanna mention before we even delve into this that this is not the whole picture. These are just the patients that we follow in audiology with CMV. I'm sure there are many, many patients that are followed at other offices with CMV, and there's many patients who maybe don't know that they have cCMV and aren't followed by any office. And so the high prevalence of hearing loss here is certainly skewed by the fact that they're being picked up in an audiology clinic. And so what I'll say is that we had 15 kids that we've identified as having CMV, and 14 of these currently have hearing loss. There is one child who has typical hearing or normal hearing and we'll be curious to see if that changes or progresses as he gets older. In eight of the kids, they had bilateral hearing loss, meaning both of their ears were impacted. In all cases the hearing loss was sensory neural. And that's not unsurprising just based on the pathophysiology of this condition. There's an entire lecture that will discuss that and I encourage you to look into that if you're interested. There was one child who had a mixed hearing loss and I'll mention that we don't believe that to be due to the CMV. He actually had a middle ear malformation that would suggest the conductive component on that side. What I think is really interesting when we look back at these patients is that I would say about half of them had severe, profound hearing loss.

And that is what we see in the literature, that severe and profound hearing loss can occur in a lot of these kids in at least one ear. And so this is really in line with what we know about CMV. It's unsurprising to us. And then in these last two boxes, you'll see the age at cCMV diagnosis, and the age at hearing loss diagnosis. For these, we are a little bit limited. Arizona is a melting pot. Lots of people come to Arizona, most likely 'cause of the weather. And in doing so, we get a lot of transfer patients, patients where records are not complete. Sometimes it'll say that the patient had cCMV, but we don't know when it was diagnosed. We don't know when the hearing loss was diagnosed. And so that's why you'll see a lot of patients that are listed as unknown for the age at

CMV diagnosis and the age at hearing loss diagnosis. But what's interesting about these first four kids under the age at CMV diagnosis is that one was identified in utero and three were identified within the first week after birth. Now, for these four kids, what's excellent is they were tested early and we know that this CMV infection is likely congenital, meaning it was acquired in utero. After that three week period, we were unable to determine if it was a congenital infection or if it was acquired later after the child was born. For the age at hearing loss diagnosis, you'll see that many of them were identified early in the first year of life, but some of them we we actually don't know. And so my takeaways from this is that most of these kids have hearing loss, but again, keep in mind they were already followed by an audiology department, so that's unsurprising. Almost half developed severe, profound hearing loss in at least one year. And despite this, only 20% of these kids were identified within the first month of life for having hearing loss.

So what's important to take away is that we're testing these kids early on and then we're testing them routinely, because hearing loss of course can develop later. Audiology interventions. The most common audiology intervention that we'll see with cCMV and hearing loss is of course the hearing aid. Even for kids who later receive a cochlear implant, it's recommended that they have a trial with a well fit hearing aid. And again, as an audiologist, it's important, it's our job, in fact to counsel families on the risk of progressive hearing loss. We want them to be selecting devices that are gonna grow with their children. And if that's not possible, we could always do loaner devices so that if the hearing loss does progress or change, the families are not financially burdened with purchasing new devices again. We're seeing more cochlear implantation for a single-sided deafness After the FDA criteria has changed. And I think for kids who have cCMV, there's another thing to think about. And that is that when hearing loss is congenital, the other ear may develop hearing loss by the time the child is 18. And so if that happens in a child with single-sided deafness and cCMV, some options may be better suited to that child than others. And what I'm thinking about

specifically is a BAHA or a CROS device which is gonna rely on that child's better ear. And if that better ear changes or progresses, that child may need additional interventions. So knowing this, families might opt for a cochlear implant earlier in life. And educating the families on this is of course important. Some children may require other accommodations or other interventions, including an FM system, assistive technology, or rehabilitation or potentially even speech and language therapies. And with that, I'd like to turn it over to Dr. Deborah Flynn, who is also an audiologist here at Phoenix Children's Hospital. She's gonna be talking to you about the prevalence of cCMV and then she's actually gonna talk about some of these patients that we follow at Phoenix Children's so that you can see the degrees of hearing loss, the configurations and how they were treated and managed here at our clinic.

- All right, thank you, Alissa. Good afternoon, thank you for joining us today in our discussion on CMV and audiology. I'm Deb Flynn. I'm gonna switch gears here and provide some case studies for children from our cohorts and describe some of the ways that we've been managing them. So I just put this slide in here as a quick reference guide for all of the statistics that we've had in our previous slides. Not necessarily gonna go through it, but I just wanted something to kind of quickly grab your attention and that way you can refer back over time. So let's jump into our first case study here.

The cases that we have, I'm gonna provide some different age ranges, different types and degrees of hearing loss, and then various treatment options. So our first kiddo is an eight year old female. She had a NICU stay of about three months and while she was in the NICU, she was diagnosed with cCMV. She passed her newborn hearing screening, went home and developed age appropriately. Her speech and language also developed age appropriately. And she passed a hearing screening when she got to kindergarten. Between kindergarten and first grade, her mom and dad started noticing that she was asking for frequent repetition and her teacher had concerns that she

wasn't able to follow directions consistently. Now this family lives in a fairly rural area about three hours from our clinic. And so they referred her over to an ENT who primarily provided services to adults, and they even had to travel to that ENT. Once she was assessed through the ENT, the audiologist found that she had hearing loss. So she had asymmetric hearing loss and they fit her with a RIC hearing aid because that's really all they had at the time, with a dome until she could get down to our clinic. So let me switch here. This is her audiogram. We repeated the audiogram when she got to our clinic and it was actually very similar to what they found at the ENT'S office where she originally went.

So she had a moderately severe rising to moderate hearing loss in her right ear, and a profound hearing loss in her left ear. I did a little aid of testing with her RIC hearing aid just to kind of get an idea. And it was as expected for a RIC hearing aid with a open dome. So of note, her right ear, her better ear was only responding with a 68% discrim. So she was really struggling at this point. We got her fit with BTE hearing aids and evaluated for a cochlear implant in her left ear. Her family was very eager to move forward with anything that would help her, because she was struggling. So she was implanted in May and she made excellent progress with her cochlear implant. She returned for her two month followup and had open-set speech with her cochlear implant. While I removed her cochlear implant, she was wearing her hearing aid. I noticed that she wasn't responding very well and was really struggling. So thought it was her hearing aid. I went and checked her hearing aid and it was fine, put her back in the booth. And this is only two months after her cochlear implant and her right ear had already progressed.

So now her better ear was in the moderate sloping to profound range. So you can see that she's doing quite well with her aided detection from her cochlear implants. But her aided benefit was pretty poor. So just to make sure that this was not a fluctuation in hearing, I brought her back about a month later and we confirmed that it was indeed a

primarily sensory neural hearing loss. And her decrim dropped from 68 to 24. So her right ear was really not doing much for her. So I had a conversation with Mom and I said, let's think about a cochlear implant for that right ear. Mom was a little concerned because she had been so dependent on the right ear. She was starting a new school year and the family lived six hours away. They just finished their activation series for the first year, which means that they would have to go through the activation series for the second year. In the end, the family decided to proceed and she was implanted in September of 2022. This was five months post activation. And they came back and she was doing quite well for this little tiny audiogram, sorry, with her bilateral cochlear implants. And her discrimination improved to 84%. Her HINT sentences was a hundred percent and her speech and noise was actually up into the 90 percentile. So she was doing wonderful with her bilateral implants.

As you can see, she did lose the residual hearing in her right ear, but the difference between 24% and 84% is pretty significant. All right, I put in these. So I also test the difference between her cochlear implant with her right and her left ear. Remember her left ear was implanted first, and typically kids do better with their first implanted ear. But if you look at her discrimination scores for the left ear, she was at 52%. You look at her scores in her right ear and she was at 72%. So I feel like the difference between the two was that she did have a little bit of residual hearing on that right side. And we caught that change pretty quickly, where the left ear, we don't know how long she had that profound hearing loss. So she really didn't come to us until she had already been diagnosed with a profound hearing loss and we were not sure the timeframe. So again, the earlier the better for consideration of cochlear implants. All right, let's talk about case two.

Case two is a three year old male. He had a very complicated birth history with cardiac arrest at birth, HIE in the basal ganglian area, which also controls movement of his muscles. And this was all due to Group B Strep sepsis. He had neonatal seizures and

he had hyperbilirubinemia. While he was in the NICU, he was diagnosed with cCMV virus. He failed his newborn hearing screening and was seen for a diagnostic test at 21 days. And rather than the typical sensory neural hearing loss, he was diagnosed with ANSD. So let me show you his waveforms here. So it was a little noisy 'cause we were in a NICU room. But overall results are showing ANSD. He has been monitored over time. So we did some behavioral testing with him via audiogram. And overall he was actually responding quite well until most recently.

So remember, he's three. His most recent audiogram showed a mild hearing loss in the left ear and a moderate hearing loss in the right ear. He has type A tympanograms and absent acoustic reflexes, which is consistent with ANSD. So this is his current audiogram. As you can see, he's got those emissions present, he's got normal tympanometry and just a mild hearing loss in the right ear. So we will be considering hearing aids for that right side and trying loaner devices. So his parents feel that he actually responds quite well. He has awareness of his name, he can respond. He has awareness of environmental sounds and he is developing speech and language overall except for some articulation errors. So we're gonna monitor him closely and do a trial with hearing aid.

All right, case three is a five year old female who was born full term with a healthy pregnancy. And interestingly, I'm not sure how she got diagnosed with CME because she doesn't have the comorbidities that some of the other kiddos have. But she was diagnosed and she had a newborn hearing screening, passed in her left ear and failed in her right ear on the first attempt. They repeated testing and she passed on both sides. The good news was that her pediatrician knew that it was important to refer for diagnostic testing and she was seen at 13 days for her BAER study. So her BAER study showed just a very mild hearing loss in the right ear and a normal hearing in the left ear. Her otoacoustic missions were absent in the right ear and present in the left ear. So we knew something was going on in that right side. This is just her test results.

So she did not return for about a year. So despite recommendations to kind of come back and periodically repeat testing, we got her back in a year and they did some behavioral testing and they couldn't rule out hearing loss, so a BAER study was recommended. And when they did the BAER study it was under anesthesia. So she had normal hearing in the left ear and severe to profound hearing loss in the right ear.

As you can see, so even at this time, despite the fact that her left ear is normal, her emissions are not great on that left side, completely absent on the right side, which we'd expect. And then she had tubes. So she had patent tubes on the day of testing. All right, so this was quite a few years ago, and cochlear implants were not FDA approved for SSD at the time. And so we gave the family options of treatment. Two options of course are the BAHA and the CROS, but due to her age the BAHA was chosen and selected and she did fantastic with her BAHA. She was actually very good about wearing her BAHA, and she attended an oral learning school here in Phoenix and did quite well. She returns consistently now for her monitoring, and because she's five, her family has asked to consider a cochlear implant for that right ear. So she is currently undergoing a workup and actually will be implanted this month. All right.

And our last case study is a six year old male with a complex medical history, including bilateral hearing loss, global developmental delay, cCMV, and autism spectral disorder. So he was actually born in Florida, diagnosed in Florida and initially treated before he moved to Arizona. He had chronic otitis media and multiple sets of tubes. So his family reported that he failed the newborn hearing screening. He was diagnosed with hearing loss in both ears, the right greater than the left, and the right ear was progressive. So as it progressed they fit him with a hearing aid. So when he came to Arizona, he had a hearing aid in the right ear and nothing in the left ear. So there was not a lot of history and the family didn't have results to transfer over. So we did follow up the BAER study in Arizona and this is what we found. So at the time he had a patent tube in the left ear and he had a moderate rising to mild conductive hearing loss. And then of course the

right ear had dropped to the profound range. So, family was very interested in helping him in any way, because again, he has autism.

And so his language was already delayed from that and they wanted to make sure that they were able to provide him with the best hearing opportunities, and they chose to move forward with a cochlear implant. So he was implanted in his right ear in August of 2021. He came back at a year post activation and it was quite difficult getting testing because he was autistic. But this was our first attempt, and his cochlear implant, he was doing quite well. His hearing aid, he was doing great, and the left ear was still kind of in that moderate range, but I noticed that he had normal tympanograms. So his eardrum had healed over, the tube had extracted, he healed over and he still had a moderate hearing loss. Hmm. He wouldn't do any follow-up testing at that day, he was done. So I brought him back and we found that his left ear had progressed to sensory neural. At this point, hasn't changed his treatment, he still wears a hearing aid on that side and he has the cochlear implant on the right side. So I did do some speech testing. You can see his cochlear implant he's at 60% and his hearing aid he's at 68%. I actually feel it's probably better than this, but because of the language delay from his autism, this may be kind of lower than what we'd expect. So we are monitoring him very closely. His mom is on top of it and if she has any concerns, she's great about giving us a call and coming in for testing to make sure that nothing has changed.

All right, so as previously mentioned, I'm not gonna go over this, but just think about being flexible in your treatment options and making sure that you offer the families all of the different types of treatment possible. So ACIA, which is the American Cochlear Implant Alliance, has presented guidelines now for cochlear implant candidacy for pediatrics, 'cause the FDA criteria is pretty strict. So these are some of the things that you can kind of consider while you're determining what you'd like to do for treatment options. If the audiometric thresholds are greater than 70 db, if word discrimination is less than 50%, or they are performing very poorly or making limited progress, then it's

important to consider cochlear implants. So if you're at a clinic that doesn't do cochlear implants, consider referring out. If you are at a center that has cochlear implants, then make sure you get them over to the cochlear implant team for at least an evaluation and monitoring.

All right, so some of the take home messages is that most states don't have the universal newborn hearing screening, but this is the time I feel like we're starting to get awareness and it's important that we begin the process of trying to get this passed in each state. CMV is not normal, but it is gaining awareness, and with improvements in testing options and the abilities to do it more cost effectively, I think universal screening is becoming more feasible. So now is the time to act, and I encourage you to start advocating in your home state. If you know of a group working on feasibility testing, consider joining and working for the fight for universal screening. So just as universal newborn hearing screening changed the way that we practiced audiology about 20 to 25 years ago, I think that universal CMV testing is also going to further change our understanding and our treatment of progressive hearing loss. I think that the statistics that we presented today are probably an underestimate of the actual children born with CMV and of those kids that we see with progressive hearing loss in our clinics. So my hope with the Universal Newborn Hearing screening, we're better able to monitor for progressive hearing loss and treat hearing loss earlier with the intent to make the lives of these children with CMV better.

- That is fabulous. All righty, well, we have some time for some questions, so please put your questions in the Q and A spot. We have a couple right now. For Deb and Alissa, what do you say to parents? How do you form the question, if the parent doesn't know what the cause of the hearing loss is and you see symptoms of CMV, what do you say to parents? Do you encourage them to get tested or bring it up, or what goes through your mind and how do you talk to parents?

- Yeah, I'll take that. So I think it's so important to be talking to these families, especially when if they've had a genetic panel and it's come back normal and we don't know what's causing the hearing loss, I think we need to be on high alert that a patient could have cCMV. One of the things that we know is that when women are educated about the risk of contracting CMV in utero, fewer children contract CMV. And so even if it was too late for this child who may have CMV, even just educating this parent might help other families. But for this child specifically, it's important that we educate them, tell them that it's possible that it could be related to cCMV, have them talk to their pediatrician. You could always test them depending on how old they are, but of course if it's after three weeks of age, it's gonna be unclear if that was a congenital infection. But families need to know that this is a possibility because it could be progressive, it could have major implications on what devices they use in the future. And I think education is really the first step for treatment. And also it's the first step for getting universal screenings for CMV in our country. The more people that know about it, the better. And I think every opportunity with a patient like this is a chance to educate.

- That makes a lot of sense. I have a follow-up question to that. Do you counsel, I'm just thinking about counseling for treatment. If you don't know the cause of the hearing loss, if you don't know if it's CMV or something else, what do you say to parents when they want those cute little teeny tiny, colorful aids, but you think it might be CMV and you need to counsel them for something? How do you do that?

- Yeah, well there are some papers that'll talk about some suggestions that could be CMV, so like a white matter abnormality on imaging that might be suggestive of CMV. So if you're seeing that in the chart, you're seeing these signs and symptoms of CMV, I would definitely discuss that with them. Small, cute hearing aids are fun, they're cute. People love 'em, I love them, but it's maybe not always the best for our patients. And so if we can encourage families to consider devices that might grow with them, I think everybody's better off in the long run. And so I usually walk them through that. If you

say, I don't know what's the cause of your child's hearing loss, sometimes hearing loss stays the same, sometimes it gets worse, it progresses. These are your options. And maybe hearing aids more powerful, slightly larger might be better for you and your family.

- That makes a lot of sense. We have a question from Jean. She's wondering, is ASD a common comorbidity with cCMV?

- I'm sorry, ANSD?

- Yes.

- Yes. So at least in our cohort, we only have one with ANSD, and I believe that the ANSD is more from the comorbid complications than actually from cCMV itself. So most of the time hearing loss from cCMV is sensory neural in nature. So I think that was just kind of a unusual one, but I wanted to present it because it was so unusual. And I think at this point, because we don't have universal screening, we truly don't know what the incidence of each of the different types of hearing loss are that we're gonna have conductive hearing losses on top of, on top of sensory neural hearing loss, which results in mixed hearing losses. Or you might have kids who need tubes. So I think we'll find more once we start testing more.

- What about Autism Spectrum Disorder? Is that a common comorbidity with cCMV?

- I don't think that we have an answer yet. I think that, again, in our cohort we only have that one, but I don't think that they're at any more risk than the normal population. But it's certainly something as an audiologist that you will have to work with and find unique ways in testing. So I think that's the most challenging is monitoring hearing and if you can't get behavioral testing, you may have to opt for sedated ABRs.

- That all makes sense. We have another question about vision differences. If there's a child that cannot see, how do you assess their hearing? When they're little, how do you do that?

- That's a great question. That's a really good question. So I have a couple who have some visual impairments and when I'm using VRA, I actually turn the lights off. So I'll turn the lights off in the booth and our setup in our clinic is that I actually sit in the booth with them and test right in front of them. So I'll sit right in front, I'll have the light from the screen and I just watch their faces and do the conditioning with our lights. So it is not easy, and if we can't get anything with VRA, then we'll turn to BAER study.

- Can you give us just like, what sort of things go through your mind? What's your thought process? 'Cause this is really important just to teach people how to do this. Really, what goes through your mind? Tell me more, Deb, tell me more.

- Okay. I think with visual impairment, the other big thing to keep in mind is that we do have otoacoustic emission testing as at least a monitor. So because they have multiple comorbidities, and oftentimes they're non-ambulatory and non-verbal, I have one kiddo that we primarily use otoacoustic emmissions to monitor his better ear. So he has normal hearing in one side and a cochlear implant on the other. And so he's not able to test VRA. Anytime he has a procedure for another medical condition, Mom always calls and says, hey, we're having a test under anesthesia, can we add a ABR with it? Absolutely. So we've been able to monitor every six months and we do usually one under anesthesia and then we do his otoacoustic emmission screening.

- Yeah, that kinda speaks to the multidisciplinary approach. Many of these children, especially if they're symptomatic, they're gonna see lots of specialists, and so the

coordination that you did with that patient is probably really appreciated, so that he's only undergoing anesthesia that one time.

- Yes. I am also wondering about screening. Do you have any stories or experiences about people who have had children and they want to get their child tested for CMV? Do you have any information about that experience for moms?

- That's a great question. So if parents are suspecting CMV?

- Yes.

- That's kinda the question?

- Yeah, so if the parents are suspecting, then I encourage them to go talk to their pediatrician because yes, they can request through their pediatrician for testing. So if they happen to come in for a non sedated ABR and we find hearing loss, because we don't have a way to test yet, I tell 'em to go to their pediatrician and ask for a test.

- And I actually have some firsthand experience with that. I have a newborn at home and the pediatrician did do some CMV testing on my baby after he was born. And I think sometimes as audiologists, with cCMV being maybe not understood by some medical professionals, you may have to do some education with these healthcare providers. If your pediatrician's not familiar with it, you can provide education and that might actually help other families in the process.

- Good point. Do you find, like, thinking about the multidisciplinary process, do most other providers that you encounter know about cCMV and what's going on? Or do you have to explain it to them? Just tell me a little bit about that.

- I would say a little bit of both. So, I'm smiling because I recently educated a child life specialist at our hospital about cCMV and she ended up putting up some posters to educate her colleagues in the break room about it because she was shocked that it's the leading cause of non-genetic childhood sensory neural hearing loss and she didn't know about it. So, that's a great example of someone who's been working in healthcare for a long time and didn't know. But then I think other specialists do, especially some of the pediatricians that we work with, the otolaryngologists that we work with, of course they're aware, but there's an article actually that Kathleen Muldoon and others published that talks about knowledge of CMV from different healthcare providers. OTs, PTs, SLPs, audiologists. And globally, the picture suggests that many healthcare providers don't know. And so this is a great opportunity to take this information back to your healthcare systems.

- Great, and what do you think is just an easy, convenient first step that everyone who is listening to this message can take back to their practice and do just to make a difference?

- Wash your hands.

- There you go. Okay. How about you, Deb?

- I would say just advocate as much as possible and educate. I think that's the biggest thing is just start the conversations with families, start the conversations with your colleagues, start the conversations with the pediatricians that you work with, because as Dr. Nickerson kind of alluded to, Dr. Muldoon has found that there really is a lot of lacking knowledge in this area from the pediatricians. So we we're gonna need pressure not only from audiology, but from all of the healthcare providers that provide services to these kiddos.

- That makes a lot of sense. Okay, with that being said, are there any more questions from the group? If they are, put 'em in the chat. If not, we can be done. Thank you, Christy. I'm gonna speak for the group and say we've all had a lovely time presenting to all of you and it's been a lot of fun.

- [Christy] Thank you so much Dr. Steuerwald, thank you so much to Dr. Flynn and also Dr. Nickerson. I really appreciate it. We hope you all enjoyed this course, and please , if you miss the other courses in the series, they are available to you on demand.
Everyone have a great day.

- Thank you.

- [Deborah] Thank you.